

Appln No.: 09/639,207  
Applicant(s): Parsa Kazemi-Esfarjani et al.  
AN ANIMAL MODEL OF POLYGLUTAMINE TOXICITY,  
METHODS OF USE, AND MODULATORS OF  
POLYGLUTAMINE TOXICITY

### **20CAGHA**

CTGCAGGCCAGCGTCCTGATAAGTGAATTTCGCCGCCACCATGGGAG  
GCCCACCGTCAACCCCCCAGCAGCAGCAACAGCAGCAGCAACAGCA  
ACAGCAGCAGCAACAACAGCAGCAGCAACAGACTAGTCGTACGTAT  
CCCTATGACGTGCCCCGACTATGCGTAG

### **127CAGHA**

CTGCAGGCCAGCGTCCTGATAAGTGAATTTCGCCGCCACCATGGGAG  
GCCCACCGTCAACCCCCCAGCAGCAGCAACAGCAGCAGCAACAGCA  
ACAGCAGCAGCAACAACAGCAGCAGCAACAGCAACAGCAGCAGCAA  
CAACAGCAGCAGCAACAGCAACAGCAGCAGCAACAGCAGCAGCAAC  
AGCAACAGCAGCAGCAACAACAGCAGCAGCAACAGCAACAGCAGCA  
GCAACAACAGCAACAACAACAGCAACAGCAGCAGCAACAGCAGCAG  
CAACAGCAACAGCAGCAGCAACAACAGCAGCAGCAACAGCAACAGC  
AGCAGCAACAACAGCAGCAGCAACAGCAACAGCAGCAGCAACAGCA  
GCAGCAACAGCAACAGCAGCAGCAACAACAGCAGCTGCAACAGCAA  
CAGCAGCAGCAACAACAGCAGCAGCAACAGACTAGTCGTACGTATC  
CCTATGACGTGCCCCGACTATGCGTAG

**FIG. 1A**

### **20QHA**

MGGPPSTPQ<sub>20</sub>TSRTYPYDVDPDYA

### **127QHA**

MGGPPSTPQ<sub>127</sub>TSRTYPYDVDPDYA

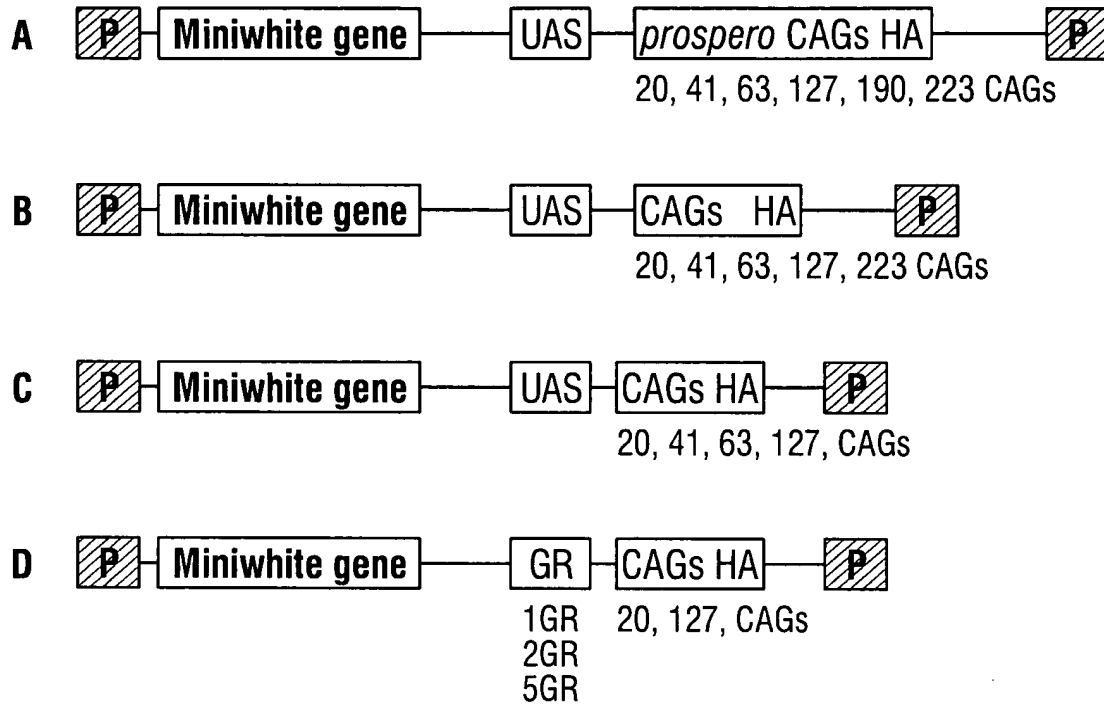
**FIG. 1B**



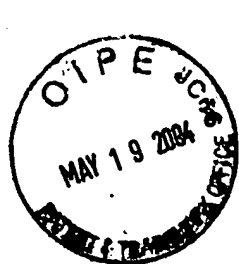
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**FIG. 2**



**Generation of P-element insertion and screening for modifiers**

**M** P[Δ2-3]/P[Δ2-3] **X F** EP55/EP55

↓

**M** EP55/Y;;P[Δ2-3]/+ **X F** w/w

↓

**M** w/Y;pEP/+;+ **or** w/Y;+;pEP/+ **X F** w;GMR/CyO;127Q/127Q

↓

Progeny screened for eye phenotype

**Isolation of the new P-element insertion (pEP=suppressor or enhancer)**

**M** (GMR;127Q)/pEP **X F** (CyO;TM3)/Xa

↓

**M** GMR/CyO;pEP/TM3 **X F** w1118

↓

**M** GMR;TM3 **or** CyO;pEP **X F** w;GMR/CyO;127Q/127Q to test

**X F** (CyO;TM3)/Xa to establish line

↓

**M** +/CyO;pEP/TM3 **X F** +/CyO;pEP/TM3

↓

pEP/TM3 **or** pEP/pEP established lines

**FIG. 3**